Cystic Replacement of Pancreas in Patient With Von Hippel-Lindau Syndrome

Varun Kapur, Steven T. Brower
Division of Surgical Oncology
Section of HPB Surgery
Department of Surgery
Beth Israel Medical Center and Continuum Cancer Centers of New York
New York, NY

CASE REPORT

A 27-year-old pregnant woman presented to our clinic with complaints of abdominal fullness, occasional nausea, vomiting, and abdominal mass. During routine ultrasound for pregnancy, the patient was found to have near-total replacement of the pancreas by multiple cysts. A fine-needle aspiration for evaluation of the mass was consistent with serous microcystic adenoma. Magnetic resonance imaging (MRI; Figure 1) revealed diffuse enlargement and replacement of the pancreas with complex cystic changes. Numerous hepatic cysts and several renal cysts were also noted. Symptoms continued postpartum, at which point a contrast-enhanced computed tomographic (CT) scan of the abdomen (Figure 2) was performed. The pancreatic cysts were noted to be unchanged from her previous MRI. Her medical history was significant for hepatitis B–induced chronic hepatitis, and she had undergone appendectomy in the past. Her family history was unremarkable. Of note, her blood group was O+. The molecular markers for malignancy (CEA, AFP, and CA 19/9) were reported to be within normal range.

All treatment options were discussed. In light of persisting symptoms and the patient's desire to bear more children, we recommended a subtotal pancreatectomy and splenectomy. Perioperatively, it was discovered that the cysts had almost completely replaced the pancreas; were strongly adherent to the spleen, splenic artery, and vein; had encased the splenic hilum; and had involved the transverse mesocolon (Figure 2). The affected portion of the mesocolon was resected, and a splenectomy was performed.

Pathology report was consistent with multiple serous cystadenomas of the pancreas. Based on these findings, the patient underwent genetic testing that revealed that she was positive for Von Hippel-Lindau (VHL) syndrome.

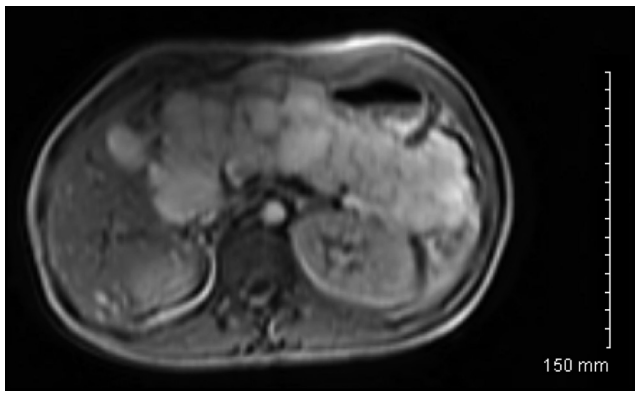

Figure 1. MRI of the abdomen.

DISCUSSION

Pancreatic lesions are commonly seen in patients diagnosed with VHL along with other manifestations (Table 1).[1] Pancreatic lesions include simple cysts, serous microcystic adenomas, adenocarcinoma, and neuroendocrine tumors.[1,2] Serous cystic neoplasms

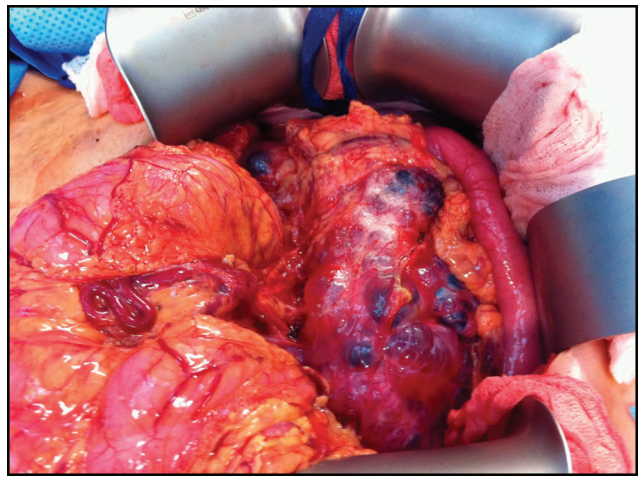

Figure 2. Perioperative finding of near cystic replacement of the pancreas.

Table 1. Manifestations of VHL disease according to prevalence

Manifestations	Prevalence (%)
Pancreatic cysts	50–91
Cerebellar hemangioblastoma	44–72
Renal cysts	59–63
Retinal hemangioblastoma	45–59
Renal cell carcinoma	24–45
Spinal cord hemangioblastoma	13–59
Pheochromocytoma	0–60
Neuroendocrine tumor of the pancreas	5–17
Serous cystadenoma of the pancreas	12
Medullary hemangioblastoma	5
Papillary cystadenoma of the epididymis in men	10–60

(SCNs) are present in approximately 10% to 12% of VHL patients.[2–5] However, as discussed in a recent review of literature by Charlesworth et al,[2] pancreatic manifestations of VHL disease are rarely the primary presenting tumors and seldom invoke diagnostic suspicion. Patients are generally asymptomatic or present with mild, nonspecific symptoms.

Ozkurt et al[6] published a case series of five patients who were hospitalized with long-term abdominal distention and pain. On examination, all had abdominal tenderness with no other accompanying findings. On imaging, all had multiple cystic lesions of the pancreas. VHL was diagnosed in all the patients, with only one having a significant family history. Girelli et al[7] also submitted a case series of five patients who presented with polycystic pancreas as the main or the only manifestation of disease and were later diagnosed with VHL. Near complete replacement of the pancreas, however, is an unusual finding. Most congenital pancreatic cysts are multiple, and almost all are associated with underlying congenital diseases. Apart from VHL, cystic fibrosis and autosomal-dominant polycystic kidney disease are also implicated.[7–10] It is important to note that cystic fibrosis is not associated with cystic neoplasm of the pancreas.[9]

Of interest, pancreatic lesions in VHL may precede other manifestations of the disease by several years[2,9,11] and can thus permit earlier diagnosis in patients with a significant family history. Recently, Weisbrod et al[12] observed a trend toward an association between pancreatic disease manifestation in VHL, especially with neuroendocrine tumor, in patients in blood group O. Our patient, as mentioned, was O+. The authors noted statistical limitations in their study, and further research is necessary before any conclusions can be drawn.

As discussed in prior studies, we agree that for all patients who present with multiple pancreatic cysts, testing for von Hippel-Lindau syndrome should be included as part of the diagnostic workup.

REFERENCES

1. Leung R, Biswas S, Duncan M, et al: Imaging features of Von Hippel-Lindau disease. *Radiographics* 28:65–79, 2008
2. Charlesworth M, Verbeke CS, Falk GA, et al: Pancreatic lesions in von Hippel-Lindau Disease?—a systematic review and meta-synthesis of the literature. *J Gastrointest Surg* 16:1422–1428, 2012
3. Kobayashi N, Sato T, Kato S, et al: Imaging findings of pancreatic cystic lesions in von Hippel-Lindau Disease. *Intern Med* 51:1301–1307, 2012
4. Taouli B, Ghouadni M, Corréas JM, et al: Spectrum of abdominal imaging findings in von Hippel Lindau disease. *AJR Am J Roentgenol* 181:1049–1054, 2003
5. Lee KH, Lee JS, Kim BJ, et al: Pancreatic involvement in Korean patients with von Hippel-Lindau disease. *J Gastroenterol* 44:447–452, 2009
6. Ozkurt H, Degirmenci H, Yenice G, et al: Huge pancreas helps diagnosis in von Hippel-Lindau disease. *Indian J Gastroenterol* 25:90–91, 2006
7. Girelli R, Bassi C, Falconi M, et al: Pancreatic systic manifestation in von Hippel-Lindau disease. *Int J Pancreatol* 22:101–109, 1997
8. Berrocal T, Pajares MP, Zubillaga AF: Pancreaticcystosis in children and young adults with cystic fibrosis: sonographic, CT, and MRI findings. *AJR Am J Roentgenol* 184:1305–1309, 2005
9. Demos T, Posniak H, Harmath C, et al: Cystic lesions of the pancreas. *AJR Am J Roentgenol* 179:1375–1388, 2002
10. Agrons GA, Corse WR, Markowitz RI, et al: Gastrointestinal manifestations of cystic fibrosis: radiologic-pathologic correlation. *Radiographics* 16:871–893, 1996
11. Hough DM, Stephens DH, Johnson CD, et al: Pancreatic lesions in von Hippel-Landau disease: prevalence, clinical significance and CT findings. *AJR Am J Rotentgenol* 162:1091–1094, 1994
12. Weisbrod AB, Liewehr DJ, Steinberg SM, et al: Association of type O blood with pancreatic neuroendocrine tumors in Von Hippel-Lindau syndrome. *Ann Surg Oncol* 19:2054–2059, 2012

Disclosures of Potential Conflicts of Interest

The authors indicated no potential conflicts of interest.

Address correspondence to: Steven T. Brower, Division of Surgical Oncology, Section of HPB Surgery, Department of Surgery, Beth Israel Medical Center and Continuum Cancer Centers of New York, First Avenue and 16th St., New York, NY 10003. Phone: 212-420-4335; Fax: 212-844-7696; E-mail: sbrower@chpnet.org.